Spontaneous pneumothorax
Pneumotorace spontaneo

Heinrich Matthys
Medical Director emeritus, University Hospital, Freiburg, Germany

Pneumothorax is defined as the presence of air in the pleural space due to: a) communication between alveolar spaces and pleura; b) direct or indirect exchange of air between the atmosphere and the pleural space; or c) the presence of gas producing organisms in the pleural space.

From the clinical and etiological standpoint the pneumothorax is classified as: **primary spontaneous pneumothorax** if occurring without obvious reason or apparent lung disease, **secondary spontaneous pneumothorax** if due to a well known underlying lung or systemic disease, or as **traumatic pneumothorax** if it is the result of iatrogenic or non-iatrogenic blunt and/or penetrating chest interventions and injuries.

Primary spontaneous pneumothorax (PSP) is therefore defined as the presence of air in the pleural space without apparent underlying lung disease or trauma. The pathogenesis of PSP is not the same for all events. Most authors believe that the communication of air between the alveolar spaces and the pleura is due to a rupture of subpleural blebs or bullae [1].

Although most children [2] and adults (3] present blebs or bullae, it is unclear how often this pathology is responsible for the leakage of air from the alveolar into the pleural space [4].

During thoracoscopy or surgery often there are other lesions present, such as inflammatory elastofibrotic layers with increased porosity and areas of disrupted mesothelial cells at the visceral pleura, allowing air leakage into the pleural space [5,6].

Bullectomy has a recurrence rate of up to 20% without pleurodesis, which may be explained by factors like peripheral airway inflammation due to noxious agents, e.g. tobacco smoke [7], or exposure to high levels of ozone as discussed by Abul et al. in this issue of *Multidisciplinary Respiratory Medicine* (pp. 16-19). Hereditary factors [8], anatomical abnormalities of the bronchial tree, ischemia at the apices of the lungs [9], low body mass index due to anorexia and other causes of food restriction [10], Marfan syndrome [11] as well as increased aluminium plasma concentrations [12] may also lead to abnormal connective tissue formations (fibrillopathies) predisposing for the occurrence of PSP [13].

Secondary spontaneous pneumothorax (SSP) is defined as the presence of air in the pleural space as a consequence of clinically apparent underlying lung disease without iatrogenic or other external blunt (barotraumata, etc.) or penetrating chest injuries. Frequent or typical causes of SSP are:

- chronic airway and alveolar diseases: e.g. severe asthma, cystic fibrosis, emphysema, bullae and cysts;
- infectious lung diseases: e.g. pneumonia (*Pneumocystis carinii*), TB, lung abscess leading to pneumothorax with pleural empyema;
- interstitial lung diseases: e.g. idiopathic fibrosing alveolitis, sarcoidosis, histiocytosis X, lymphangioleiomyomatosis;
- systemic connective tissue diseases: e.g. rheumatoid arthritis, ankylosing spondylitis, scleroderma, Marfan- and Ehlers Danlos-syndrome;
- malignant lung and chest diseases: e.g. bronchial cancer, sarcoma.

SSP has a higher recurrence rate than PSP, up to 80% in cystic fibrosis.

The age-adjusted incidence of PSP is 7.4-18 cases per 100,000 population each year in males and 1.2-6 in females and it occurs typically at rest. A 2nd World War study showed no difference of incidence between royal air force pilots and army soldiers. Therefore, avoiding physical stress and exercise should not be recommended. In contrast loud music as an atmospheric pressure change may be a risk factor for PSP [14]. Tension pneumothorax in PSP is rare. The outpatient treatment for the first event consists in introducing a small chest tube with a one-way valve (Matthys Drain®) followed by pleurodesis in patients at risk for recurrent PSP [15].

Heinrich Matthys
Department of Pneumology, University Hospital Freiburg,
Hochrüttestr. 17, D-79117, Freiburg, Germany
email: hmatthys@t-online.de

Multidisciplinary Respiratory Medicine 2011; 6(1): 6-7

References

1. Bradley M, Willams C, Walshaw MJ. The value of routine expiratory chest films in the diagnosis of pneumothorax. Arch Emerg Med 1991;8:115-116.
2. Guimaraes CV, Donnely LF, Warner BW. CT findings for blebs and bullae in children with spontaneous pneumothorax and comparison with findings in normal age-matched controls. Pediatr Radiol 2007;37:879-884.
3. Amjadi K, Alvarez GG, Vanderhelst E, Velkeniers B, Lam M, Noppen M. The prevalence of blebs and bullae among young healthy adults: a thoracoscopic investigation. Chest 2007;132:1140-1145.
4. Noppen M. Con: Blebs are not the cause of primary spontaneous pneumothorax. J Bronchol 2002;9:319-325.
5. Masshoff W, Höfer W. Zur Pathologie des sogenannten idiopathischen Spontanpneumothorax. Dtsch Med Wochenschr 1973;98:801-805.
6. Radomsky J, Becker HP, Hartel W. Pleuraporosität beim idiopathischen Spontanpneumothorax. Pneumologie 1989;43:250-253.
7. Bense L, Lewander R, Eklund G, Hedenstierna G, Wiman LG. Nonsmoking, non-alpha 1-antitrypsin deficiency-induced emphysema in nonsmokers with healed spontaneous pneumothorax, identified by computed tomography of the lungs. Chest 1993;103:433-438.
8. Morrison PJ, Lowry RC, Nevin NC. Familial primary spontaneous pneumothorax consistent with true autosomal dominat inheritance. Thorax 1998;53:151-152.
9. Withers JN, Fishback ME, Kiehl PV, Hannon JL. Spontaneous pneumothorax. Suggested etiology and comparison of treatment methods. Am J Surg 1964;108:772-776.
10. Coxson HO, Chan IH, Mayo JR, Hlynsky J, Nakano Y, Birmingham CL. Early emphysema in patients with anorexia nervosa. Am J Respir Crit Care Med 2004;170:748-752.
11. Neptune ER, Frischmeyer PA, Arking DE, Myers L, Bunton TE, Gayraud B, Ramirez F, Sakai LY, Dietz HC. Dysregulation of TGF-beta activation contributes to pathogenesis in Marfan syndrome. Nat Genet 2003;33:407-411.
12. Leo F, Venissac N, Drici MD, Mouroux J. Aluminium and spontaneous pneumothorax. A suggestive but unconfirmed hypothesis. Interact Cardiovasc Thorac Surg 2005;4:21-22.
13. Loeys BL, Matthys DM, De Paepe AM. Genetic fibrillinopathies: new insights in molecular diagnosis and clinical management. Acta Clin Belg 2003;58:3-11.
14. Noppen M, Verbanck S, Harvey J, Van Herreweghe R, W, Vincken W, Paiva M. Music: a new cause of primary spontaneous pneumothorax. Thorax 2004;59:722-724.
15. Matthys H. Emergency management of pneumothorax – a case report and guidelines. Multidisciplinary Respiratory Medicine 2006;3:64-66.